showed that the patient in fact had spasmodic torticollis which we think was the cause of the rotary subluxation at C1/C2.

A 37 year old man presented in November 1997 with a history of an insidious onset of progressive deformity of the neck associated with spasm of the left sternocleidomastoid and trapezius muscles and occipital pain from April of that year. He had had mild neck pain for several years. The occipital pain was left sided and became increasingly severe. Electromyography was not performed. He had. been off work for 6 months and found it increasingly difficult to sleep in a comfortable position. The only relevant history was one of "anxiety attacks".

On examination there were no signs of rheumatoid arthritis. The patient had a classic "cock robin " deformity with his head tilting to the left and turning to the right. This was associated with spasm and tenderness, but no obvious hypertrophy, of the left sternocleidomastoid and trapezius which was thought to be voluntary as it subsided when he was relaxed. Plain radiography of the atlantoaxial region was essentially normal and thus CT was obtained under a general anaesthetic as the patient was extremely anxious. After the induction of the general anaesthetic, the element of spasm and tilting of his neck which was thought to be due to psychological overreaction disappeared and examination showed persistence of the "cock robin" deformity. Spinal CT in the neutral position confirmed a C1/C2 rotary subluxation which reduced with the head turned towards the left but was exaggerated by turning to the right. There was no history of recent neck injury, rheumatoid arthritis, or pharyngeal infection and thus a cause for the C1/C2 rotary subluxation was not apparent at that stage. In view of the severity of the pain surgical stabilisation of C1/C2 was suggested.

The patient was placed in halo-traction for a week, and further CT was performed. This showed significant improvement but not total correction of the rotary subluxation. As reduction was not total, it was decided not to perform transarticular screw fixation of C1/C2 but a posterior modified Gallie fusion was performed in the position of maximum reduction. A halo-vest was applied. Check radiography was satisfactory and the patient reported a very pleasing relief of the pain and spasm which he had preoperatively. The halo-vest was maintained for 10 weeks during which his pain and spasm had completely resolved. However, shortly after removing the halo, he had a recurrence of the pain and spasm in the left sternocleidomastoid al though the severe occipital pain was still completely relieved. His neck posturing was now variable and typical of spasmodic torticollis with no residual "cock robin" deformity. There was rotation to the right, tilt to the left, and left shoulder elevation. Check radiography showed no loss of the previous position.

At this point a diagnosis of spasmodic torticollis was made with a torticollis severity score as described by Tsui *et al* of 15 indicating moderately severe deformity.[1] Five hundred units (200 mU/ml) of botulinum toxin A (Dysport—Ipsen) were injected into the left sternocleidomastoid (250 mU) and trapezius (250 mU). The first injection itself produced a 50% reduction in pain and spasm. Three further injections of the same dose over a year produced further improvement and at latest follow up 16 months after surgery his torticollis score had fallen to eight. His posture was similar in nature but much less pronounced and variable. The occipital pain remains absent and he has achieved a solid fusion.

Atlantoaxial rotary subluxations were first described by Wortzman and Dewar in 1968[2] and further clarified by Fielding and Hawkins in 1977.[3] The maximum normal rotation of the atlantoaxial joints is 45–47 degrees. Beyond this the lateral inferior facet of the atlas rocks over the lateral superior articular facet of the axis.[3]

Rotary atlantoaxial subluxation can be caused by severe twisting injury of the neck, usually associated with violent sport and vehicular accidents.[4] It has been reported as a result of a greater than 90 degree rotation of the neck under general anaesthesia.[4 5]

In this case the common causes of C1/C2 rotary subluxation were absent and we suggest that the subluxation was caused by the spasmodic torticollis over time. Surgery and application of the halo abolished the occipital pain and the spasm was reduced, possibly due to the extra somatosensory input from the halo (a mechanical geste antagonistique). Removal of the halo was followed by recurrence of the dystonic spasm but the occipital neuralgia remained absent due to the stabilisation of the atlantoaxial complex.

In this case it was not possible to determine at what stage the rotary subluxation occurred. It is possible that the subluxation was the primary event leading to malposition of the neck and muscle spasm—a type of "post-traumatic" dystonia. However, in patients with atlantoaxial rotary subluxation, the normal neck deformity is the classic "cock-robin" deformity and activation of sternocleidomastoid and trapezius does not occur. The surgical treatment in this case resolved the "cock robin" deformity and occipital pain but the typical clinical findings of spasmodic torticollis reappeared once the halo was removed. It is most likely that the halo provided sufficient somatosensory input to inhibit the sternocleidomastoid spasm during the length of time the halo was in position. The evolution of the clinical findings with relief of occipital pain and "cockrobin" deformity followed by a more typical appearance of spasmodic torticollis strongly suggest that it was the dystonia which caused the rotary subluxation.

Spasmodic torticollis is a focal and usually idiopathic dystonia with cervical muscle spasm causing involuntary neck posturing and movement. It can occur at any age. Chemical denervation of the overactive muscles with botulinum toxin is now the usual treatment and is effective in most patients.[6]

Dystonia can cause subluxation or dislocation at various joints. For instance, the temporomandibular joints can undergo recurrent or chronic dislocation in idiopathic or tardive oromandibular dystonia.[7–9] Angelini *et al* described subluxation of the subaxial cervical spine resulting in a cervical myelopathy in a child with spastic dystonic cerebral palsy[10] and Tunkel *et al* reported cervical subluxation causing improvement in the dystonia in a patient with longstanding idiopathic torsion dystonia.[11] To our knowledge adult onset spasmodic torticollis causing and presenting as a rotary atlantoaxial subluxation has never been reported in the literature. Prolonged rotation beyond the physiological limit is likely to be the cause of this subluxation.

When atlantoaxial subluxation appears after prolonged involuntary neck posturing an underlying diagnosis of dystonia should be considered. Botulinum toxin will not resolve the subluxation, but was necessary in this case to control the underlying dystonia. External braces and collars rarely control the forceful movements of cervical dystonia, and the toxin may take some days or even weeks to work, so we recommend treatment as soon as dystonia is diagnosed. In theory botulinum toxin might enhance the effect of or interfere with recovery from acute muscle paralysing agents used in anaesthesia. However, no such reactions have been reported in 12 years of extensive experience worldwide with botulinum toxin, and therefore it is probably safe to give injections even before neck surgery. When a patient with spasmodic (variable posturing) torticollis develops a fixed and sufficiently extreme "cock robin" posture, the clinician should consider investigation by plain radiography and CT to exclude rotary subluxation, even if the muscle spasm is intermittent.

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Ataxic form of Guillain-Barré syndrome associated with anti-GD1b IgG antibody

Richter[1] proposed an ataxic variant of Guillain-Barré syndrome, in which patients have severe ataxia of the cerebellar type at the onset of Guillain-Barré syndrome but no ophthalmoplegia or severe loss of proprioceptive sense. Patients with ataxic Guillain-Barré syndrome have distal paraesthesias, areflexia, and raised CSF protein concentrations. Kusunoki *et al*[2] reported that of 149 patients who had anti-GQ1b IgG antibodies without profound weakness, five had acute self limited ataxia without ophthalmoplegia. The nosology of these patients, however, was not discussed. Of our 340 consecutive patients who had anti-GQ1b IgG, six had no external ophthalmoplegia and one had minimal external ophthalmoplegia. The clinical features of these seven anti-GQ1b-positive patients were consistent with an "ataxic form of Guillain-Barré syndrome" (Yuki *et al*, unpublished observations). Tentative diagnoses made by the primary physicians were Guillain-Barré

syndrome (n=3), atypical Miller Fisher syndrome (n=3), and acute cerebellar ataxia (n=1). Araki *et al*,[3] however, reported on a patient with Guillain-Barré syndrome who had prominent cerebellar signs. That patient had high monospecific anti-GD1b IgG antibody titre in the acute phase of the illness, but did not have anti-GQ1b IgG. These findings led us to examine whether some patients in whom acute cerebellar ataxia has been diagnosed have anti-GD1b IgG antibodies.

Serum samples were obtained from 39 patients for whom acute cerebellar ataxia or acute cerebellitis had tentatively been diagnosed. One patient who had associated anti-GQ1b IgG was excluded because the tentative diagnosis was acute cerebellar ataxia and the final one ataxic Guillain-Barré syndrome. Serum IgM or IgG antibodies to GM1, GM2, GD1a, GD1b, GT1b, GQ1b, or GQ1bα were measured by an enzyme linked immunosorbent assay as described elsewhere.[4] GQ1bα is a possible target molecule for serum antibodies from patients with sensory ataxic neuropathy.[4] Serum was considered positive when the antibody titre was 500 or more. One of the 39 patients had a high anti-GD1b IgG titre of 16 000 but carried no antibodies to the other six gangliosides. His clinical presentation is that described later. The other 38 patients had no antibodies to those gangliosides. Further study showed that serum from the patient who had anti-GD1b IgG did not react with asialo-GM1, fucosyl-GM1, GM1b, GalNAc-GM1b, GalNAc-GD1a, GD3, GT1α, GT1aα, or sulfated glucuronyl paraglo-boside.

A 55 year old man had a cough and nasal discharge that disappeared after a few days. After resolution of this illness, he noted paraesthesias in his fingers and toes (day 1) which worsened, and he developed an unsteady gait on day 3. He was apyrexial and fully conscious. Blepharoptosis was absent. Ocular movement was not limited, but smooth pursuit was saccadic. His pupils were normal, and light reflexes prompt. Neither facial nor oropharyngeal palsy was present. Limb weakness was insignificant. Deep tendon reflexes were absent. Babinski's sign was negative. Finger to nose and heel to knee tests were dysmetric and uncoordinated. Romberg's sign was negative, but standing in a tandem position was unsteady. Tandem gait was impossible, and assistance was needed to walk. Paraesthesias of the glove and stocking type were present. There was no impairment of pinprick, touch, position sense, or vibratory sensation. Autonomic nervous function was normal except for hyperhidrosis in the palmar and planter surfaces. On days 3, 4, 6, 8, and 10, he underwent immunoadsorption treatment. The neurological signs rapidly disappeared. Motor and sensory nerve conduction values were normal on days 7 and 21. Protein in CSF was 32 mg/dl on day 3, and 60 mg/dl on day 10 with normal cellularity. On day 24 he was discharged without clinical signs, but still with mild paraesthesias in the right fingers. No external ophthalmoplegia was found during the course of the illness.

Acute cerebellar ataxia had been tentatively considered, but the ataxic form of Guillain-Barré syndrome, as proposed by Richter,[1] could be diagnosed. This is important because some patients in whom acute cerebellar ataxia has been diagnosed may have ataxic Guillain-Barré syndrome, and they could benefit from undergoing established treatment for the syndrome—namely, plasmapheresis or intravenous administration of immunoglobulins. Serum samples from patients for whom acute cerebellar ataxia or acute cerebellitis is diagnosed should be tested for anti-GD1b and anti-GQ1b IgG antibodies in order not to overlook cases of ataxic Guillain-Barré syndrome.

Monospecific anti-GD1b IgG antibodies have been detected in a patient with Guillain-Barré syndrome who had prominent sensory ataxia and in a patient who had acute distal paraesthesias and areflexia after upper respiratory tract infection.[5 6] Whether the presence of monospecific anti-GD1b IgG is correlated with a particular clinical condition, therefore, is uncertain. By contrast, a patient with cerebellar ataxia and chronic polyneuropathy has been reported to have IgM M-protein to GD1b, GM1, and asialo-GM1.[7] Both the monoclonal IgM with anti-GD1b activity and murine monospecific anti-GD1b monoclonal antibody bind to the human cerebellar granular layer. The binding of anti-GD1b IgG to the cerebellar granular layer or spinocerebellar 1a fibres in the peripheral nerves may have produced the patient with cerebellar ataxia reported by Araki *et al*[3] and our present patient.

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Analysis of adenosine deaminase isoenzyme-2 (ADA_2) in cerebrospinal fluid in the diagnosis of tuberculous meningitis

The outcome of tuberculous meningitis is influenced by the stage of disease at the start of treatment. Initiation of antituberculous therapy is often delayed because of the inadequacy of presently available laboratory tests. Management of patients with possible tuberculous meningitis would thus be advanced by the development of an accurate, reliable, and rapid diagnostic test, particularly if it could be applied in settings with poor recourses.

Adenosine deaminase (ADA), an enzyme involved in purine catabolism, exists in at least three forms. ADA_1 is a monomeric protein with a molecular mass of approximately 35 kDa and two ADA_1 molecules joined via a connecting protein form the dimeric ADA_{1+CP}. The third isoenzyme ADA_2 seems to be produced only by monocytes.[1] Total CSF ADA has been suggested as a marker for tuberculous meningitis[2 3]; however, considerable variability and overlap, particularly with acute bacterial meningitis, has led some authors to question its clinical usefulness.[4]

An increased proportion of ADA_2 has been suggested to be a more specific means of diagnosing tuberculous effusions.[2] However, the use of CSF ADA_2 in the diagnosis of tuberculous meningitis has not been described.

Study subjects (all adults) were from a prospective cohort of patients undergoing a diagnostic lumbar puncture for suspected meningitis. Patient characteristics and investigations have previously been described in detail.[5] In addition, CSF specimens from 10 patients at the Johannesburg Hospital were included (four tuberculous meningitis, four cryptococcal meningitis, two acute bacterial meningitis). Serum and CSF ADA analysis was performed on 11 specimens from patients with tuberculous meningitis (nine established by culture, two probable), nine with cryptococcal meningitis, 13 with acute bacterial meningitis (nine *Neisseria meningitidis*; two *Streptococcus pneumoniae*), nine with aseptic meningitis, and 19 with a normal lumbar puncture. All patients with tuberculous meningitis, cryptococcal meningitis, or aseptic meningitis, six of 13 with acute bacterial meningitis, and 10 of 19 of the normal lumbar puncture groups were HIV positive. Ethical approval was obtained from the committee for research on human subjects of the University of the Witwatersrand.

Tuberculous meningitis was confirmed if the CSF culture yielded *M tuberculosis*. Probable disease was diagnosed in the presence of a lymphocytic pleocytosis (>20 cells/mm^3), high CSF protein (>0.8 g/l), low CSF glucose (<60% of matched plasma glucose), and evidence of pulmonary tuberculosis. Cryptococcal meningitis) was diagnosed if either the indian ink stain, CSF fungal culture, or CSF cryptococcal antigen were positive. Acute bacterial meningitis was diagnosed in patients with acute onset of symptoms, pyrexia, a CSF neutrophilia with high protein, and low CSF glucose. Aseptic meningitis was diagnosed if there was a predominantly lymphocytic pleocytosis, a normal or moderately raised CSF protein (>0.7 g/l), and negative serology and bacterial, fungal, and mycobacterial culture.

Analysis of CSF was considered normal when there were < 5 leucocytes/mm^3, protein <0.45 g/l, and negative culture and serology.

Matched CSF and serum samples were frozen within 6 hours of collection, stored at−20°C (the enzymes are stable for at least 4 weeks) and analysed within 1 week of collection. Tests were performed with laboratory staff unaware of the diagnosis. Total ADA activity was measured by an enzymatic spectrophotometric method on a Cobas Mira autoanalyser (Roche Diagnostics, Switzerland). Erythro-9-(2-hydroxyl-3-nonyl)-adenine, a selective ADA_1 and ADA_{1+CP} inhibitor, was added to the reaction mixture at a concentration of 200 μM, allowing for the measurement of ADA_2 activity (in the same enzymatic system). The between batch coefficient of variation of the test at a dilution of 6 U/l was 8%. The proportion of total ADA which comprised ADA_2 could only be reliably estimated when total ADA was >2 U/l. ADA_2 isoenzyme analysis is therefore only reported in the groups with a median total CSF ADA of >2 U/l.